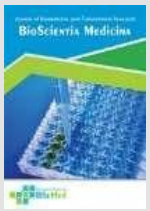
eISSN (Online): 2598-0580



Bioscientia Medicina: Journal of Biomedicine & Translational Research

Journal Homepage: www.bioscmed.com

Diagnosis and Management of Choledochal Cyst in One Year-Old Baby: A Case Report

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ARTICLE INFO

Keywords:
Biliary obstruction
Children
Choledochal cyst
Duct cysts
Icteric

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All authors have reviewed and approved the final version of the manuscript.

https://doi.org/10.37275/bsm.v7i2.775

A B S T R A C T

Background: Choledochal cysts (CC) are rare congenital anomalies of the biliary tract. The classic triad of jaundice, abdominal pain, and right upper quadrant mass are rare symptoms, mainly in children and adolescents. This study aimed to present the diagnosis and management of choledochal cyst cases in a baby girl. **Case presentation:** A 1-year-old girl, domiciled outside the city of Padang, was brought to the hospital by her parents with complaints of repeated vomiting for 1 day before being admitted to the hospital. This complaint has been accompanied by yellowish eyes and body, pale bowel movements, and dark brown urine since 3 months ago. The patient was never brought to the doctor for treatment. Since 1 month ago, the patient often feels pain in the abdomen and has a lump in the upper right abdomen. On the abdomen area, there was no distention, hepar was ¼-1/4 sharp edge, tenderness at the epicondrium area, no rebound tenderness, turgor was decreased, and no palpable mass. Abdominal ultrasonography presented a choledochal duct cyst with biliary obstruction. Abdominal CT scan with contrast revealed intrahepatic and extrahepatic biliaryectasis et causa suggestive of the choledochal cyst (Todani IVA) and hepatomegaly. The patient was consulted by the pediatric surgery division for further management and was advised to undergo surgery with the Roux En-y procedure. Follow-up after the patient underwent surgery showed clinical improvement. Symptoms of jaundice in the eyes and skin begin to decrease, as brownish stools and light brown urine. **Conclusion:** Surgical treatment with the Roux-en-Y procedure is the definitive treatment for choledochal cysts in this patient.

1. Introduction

Choledochal cysts are uncommon congenital anomalies of the biliary tract.[1,2] Approximately 80% of choledochal cysts (CC) are diagnosed in infants and young children within the first decade of life.[3] Choledochal cysts are four times more common in females. In symptomatic patients, the presentation may include jaundice, cholangitis, and pancreatitis and is typically present in infancy and childhood. Clinical presentations of CC include nonspecific abdominal pain.[4,5] The classic triad of jaundice, abdominal pain, and right upper quadrant mass are rare symptoms, mainly in children and adolescents. Clay-color stools are often found as a clinical sign of CC. The choledochal cyst will block the flow of bile, then bile flows back to the liver and enters the systemic circulation. Bilirubin will accumulate in the blood and manifest as dark brown urine, jaundice, and reduced bilirubin in the feces (pale stool).[6,7] This study aimed to present the diagnosis and management of choledochal cyst cases in a baby girl.

2. Case Presentation

A 1-year-old girl, domiciled outside the city of Padang, was brought to the hospital by her parents with complaints of repeated vomiting 1 day before being admitted to the hospital. This complaint has been accompanied by yellowish eyes and body, pale bowel movements, and dark brown urine since 3 months ago. The patient was never brought to the doctor for treatment. Since 1 month ago, the patient often feels pain in the abdomen and has a lump in the upper right abdomen. The patient was then taken to a local hospital for examination. At the examination, it was found that the patient had repeated vomiting 1 day before entering the hospital. The contents of the vomit were in the form of food and drink, non-projectile vomiting, no fever, and bleeding. The child's weight decreased from 10 to 9.4 kg. The child has performed abdominal ultrasound with the impression suggestive of a choledochal duct cyst with biliary obstruction. Abdominal CT scan evaluation with contrast was suggested intrahepatic and extrahepatal biliary ectasis et causa suggestive of duct cysts; there was the imaging of hepatomegaly, and the Tohoku score was 7. The patient was then referred to Dr. M. Djamil General Hospital for further examination.

Based on physical examination, in general, the patient appeared moderately ill, and the heart rate was 120 bpm; the respiratory rate was 22 times per minute, regular and deep breath; axillary temperature 37°C; blood pressure was 92/60 mmHg; O_2 saturation was 98% (room air). Nutrition status was well nourished. The eyes were sunken, and the sclera was icteric. On the abdomen area, there was no distention, hepar was ¼-1/4 sharp edge, tenderness at epicondrium area, no rebound tenderness, turgor was decreased, and no palpable mass. Hematologic examinations revealed Hb level 10,2 g/dl, white blood cells 20.070/mm^3, platelet 954.000/mm^3, hematocrit 33 %, differential count 0/0/9/65/22/4, calcium 10,1 mg/dl, sodium serum 136 mmol/L, potassium serum was 4,4 mmol/L, blood glucose 104 mg/dL. PT 11.5 seconds, APTT 36.3 seconds, total bilirubin 4.7, bilirubin indirect 1.4, bilirubin direct 3.3, AST 89 U/L, ALT 68 U/L, and gamma GT 1387. Urinalysis results were normal. Abdominal ultrasonography presented a choledochal duct cyst with biliary obstruction (Figure 1). Abdominal CT scan with contrast revealed intrahepatic and extrahepatic biliaryectasis et causa suggestive of the choledochal cyst (Todani IVA) and hepatomegaly (Figure 2).

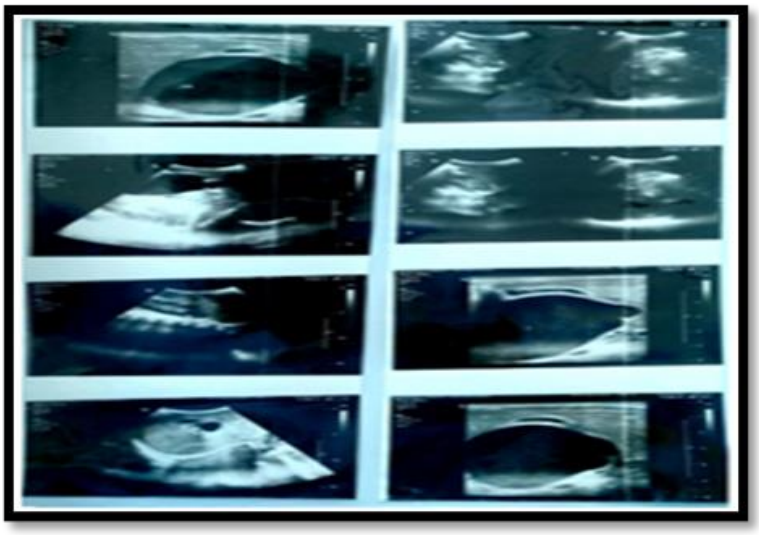

Figure 1. Abdominal ultrasound.

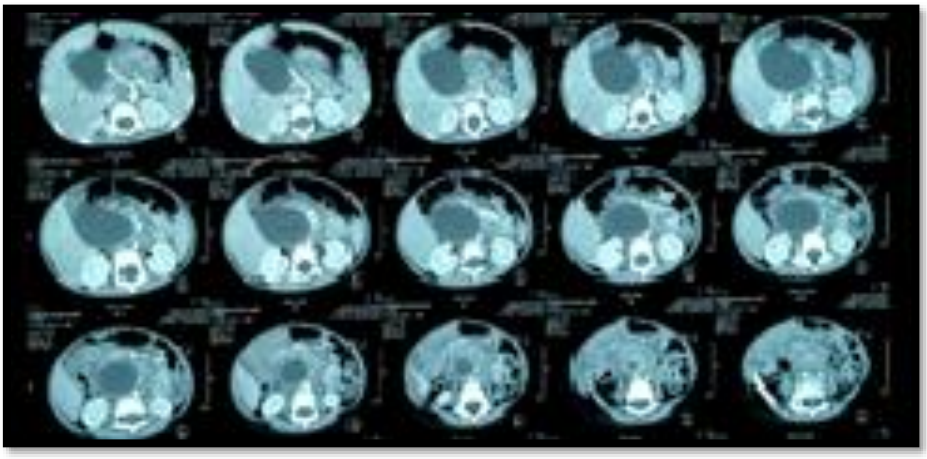

Figure 2. Abdominal CT scan with contrast.

Based on anamnesis, physical examination, and investigations, the patient was diagnosed with extrahepatic cholestasis due to choledochal biliary obstruction accompanied by vomiting with moderate dehydration. Treatment for this patient was a soft food diet of 1000 Kcal, ursodeoxycholic acid 65 mg/8 hours, vitamin A 6000 IU/day (oral), vitamin D 400 IU/day (oral), vitamin E 100 IU/day (oral), vitamins K 2.5 mg/day (oral), and paracetamol 120 mg (if the axilla temperature is more than 38.5°C). For the management of vomiting symptoms, patients are given 2A IVFD fluids 2000 mL/day and ORS 100 cc for each vomiting. The patient was consulted by the pediatric surgery division for further management and was advised to undergo surgery with the Roux En-y procedure.

Follow-up after the patient underwent surgery showed clinical improvement. Symptoms of jaundice in the eyes and skin begin to decrease, as brownish stools and light brown urine. Patients were treated with normal food consistency 1000 kcal, oral fluids 100 cc/6 hours, KAEN 1B 200 cc/day IVFD, ceftriaxone 500 mg/12 hours IVFD, penicillin 500 mg/8 hours IVFD, lansoprazole 10 mg/12 hours IVFD, ORS 100 cc/vomit, Ursodeoxycholic acid 3 x 65 mg, ursodeoxycholic acid 65 mg/8 hours, vitamin A 6000 IU/day (oral), vitamin D 400 IU/day (oral), vitamin E 100 IU/day (oral), vitamin K 2.5 mg/day (oral), and paracetamol 120 mg (if the axilla temperature is more than 38.5°C).

3. Discussion

Choledochal cysts (CC) are rare congenital anomalies of the biliary tract.[1] The incidence of CC is 1 in 150,000 individuals in Western countries to 1 in 13,000 individuals in Japan.[4,7] Choledocal cysts are classified into five types; type I: cyst of the bile duct (80% to 90% of all CC); type II: pouching or sac on the bile duct; type III: cyst within the wall of the duodenum (where the duct connects to the liver) or pancreas; type IV: dilatation intrahepatic and extrahepatic ducts (15% to 20% of all CC); and type V: multiple cysts without extrahepatic alteration or Caroli's disease.[3,8]

Clinical presentations of CC include nonspecific abdominal pain. The classic triad of jaundice, abdominal pain, and right upper quadrant mass is rare, mainly in children and adolescents.[9-12] Choledochal cysts can close the bile ducts so that bile refluxes to the liver and enters the systemic circulation. In these patients, jaundice was found in the sclera and jaundice, as well as pale feces and dark brown urine.

Multiple etiologic have been proposed for the choledochal cysts. Choledochal cysts are believed to be congenital in origin; however, the exact etiology remains unknown.[5] Choledochal cyst excision and biliary reconstruction is the treatment of choice for

choledochal cysts. Biliary reconstruction is carried out using several methods. The most popular procedure is Roux-en-Y hepaticojejunostomy.[10] The advantages of this procedure include a reduction in the incidence of anastomotic stricture, stone formation, cholangitis, and intracapsular malignancy. The clinical results of total cystectomy and Roux-en-Y hepaticojejunostomy are excellent.[8-10]

Common choledochoceles can be diagnosed at any age and found symptomatic or incidentally.[13] In addition, with modern improvements in ultrasound screening and technology, prenatal diagnosis of cysts is becoming more common. The prognosis of pediatric CC is good. In the postoperative period, early complications can include anastomotic leak, postoperative bleeding, wound infection, acute pancreatitis, and pancreatic or biliary fistula. Late complications include anastomotic stricture, cholangitis, hepatolithiasis, cirrhosis, and malignancy.[8,14]

4. Conclusion

Clinical presentations of choledochal cysts include nonspecific abdominal pain. Investigations such as ultrasound and abdominal CT scan with contrast can help establish the diagnosis. Surgical treatment with the Roux-en-Y procedure is the definitive treatment in this case.

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